TROPHIC HEMIATROPHY: COMPLETE

A TROPHONEUROSIS[1]

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I have found in literature six cases of hemiatrophy that were more or less complete, *i. e.*, that involved the face, arm and leg, or the arm and leg only, of one side. Of these the one described by Paul Broca[2] in 1859 was similar in its details to the one here presented. He said of it—a boy eleven years old—it was "as if the two halves of the body were different sized persons joined together."

In none of the references could I find any satisfactory data relative to the pathology, electrical reactions and reflexes.

Hemiatrophy of the face, including the progressive type, has often been described. Romberg is given the credit of having described the first case. Up to February, 1907,[3] 170 cases of the progressive type had been reported. Oppenheim[4] found one case in which the leg of the opposite side was involved, but at that time had never seen one in which the entire half of the body was involved—as is the condition in the case here presented. He believed the etiological factor to be "an unknown agent acting only in neuropathic individuals." Most authors believe the condition to be a trophoneurosis.

The first case found reported that resembled this one was that of Paul Broca in 1859 (vide supra). A. Stadfeldt[5] and Jacobi[6] considered, in their cases, the causal factor to have existed prior to the sixth month of fetal life. Howden[7] reported an insane woman, aged thirty years, who exhibited atrophy of the left side of the body; in this case post-mortem findings showed atrophy of the right hemisphere of the cerebrum and left side of cerebellum. In 1878[8] a case of hemiatrophy was reported and shown before the Clinical Society of London. In the same year another was shown before the Societe Anatomique by H. C. Beyer.[9] Another was reported by S. Pixley in 1883.[10] In the next year R. C. Burrell[11] reported and described a case similar to mine except that the face was not involved.

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[1] Case presented before the Clinical and Pathological Society of Los Angeles.

[2] Canslatt's Jahresbericht, 1859, Vol. IV, p. 6.

[3] Deutsche Zeitschrift für Nervenheilkunde, February, 1907.

[4] "Diseases of the Nervous System."

[5] "On the Asymmetry of the Body of the Human Skeleton."

[6] Trans. N. Y. Obst. Soc., 1871.

[7] Jour. Anat. and Physiol., Vol. IX, p. 288.

Of these six cases of hemiatrophy four showed it on the left side. Of the remaining two, I find no reference to the side involved.

I have not been able to find any case that is similar to this one reported since 1884 except one referred to in a late number of this Journal.

The points of interest attached to this case are—its rarity; the fact that the hemiatrophy is complete, *i. e.*, it involves the face, arm and leg; there is an hypertrophy of the trapezius muscle on the atrophic side; the psychic symptoms accompanying.

There are two unsatisfactory elements in it: (1) The short time that could be given to watching it because the man had to leave for the East (2) the unreliableness of a man of his type and his statements. However, the case presents much of interest, especially from a clinical standpoint.

CASE.—N. G., male, 26 years of age, unmarried. His occupations have been various: clerk, waiter, hospital orderly.

His family history is not of interest except that he has two brothers that are normal and that his mother died when he was five years old and his father when he was seven years old, when he was placed in an orphans' school.

Previous History.—He says that as a child he was not like others and would not play with them unless forced to do so. He was inclined to be solitary. Other than this he remembers nothing out of the ordinary except that during his first year at the orphans' school he began to have certain peculiar sensations; the first one came on while he was in the class room. He describes it as being like a shock accompanied by vertigo. It frightened him and he gave a cry and "staggered" to the teacher's desk. In a few moments he felt all right again. Since then there have been periods in which these "shocks" have come on daily or oftener and others when they have been absent for months. During the attack he has never been unconscious or fallen. He describes it as an electric shock that he feels all through his left side only, but without any convulsive movement whatever being visible—no muscular twitching, no biting, no involuntary micturition or the like. After the "shock" he is slightly dazed, nervous and mentally clouded: he describes it as a "fluttering." All this will last only a few minutes. The attacks have all been very much alike, a one sided psychic epilepsy, as it were.

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[8] Lancet, 1878, Vol. I, p. 387.

[9] Le Progress Médicale, 1878, p. 242.

[10] Phil. Med. News, 1883, Vol. XLIII, p. 146.

[11] Boston Med. and Surg. Journ., Vol. CXI, p. 462.

So much for the psychic element in the case.

For the last seven years he has noticed that his left side has been smaller than the right; he is not sure whether the left side has grown smaller or the right side larger, leaving the left as it was. He has evidently not been much exercised about this, as he did not volunteer any information concerning it and it was only noted by me when I stripped him for my routine examination. He says that for the last four or five years he has noticed that the left side of his neck has been larger than the right.

He does not remember having any serious illness during his life.

Present State.—He complains that the "shocks" interfere with his ability to attend to any kind of work because of the momentary weakness and mental cloudiness. He says he is in a constant state of apprehension and complains of inability to sleep. He says he is easily fatigued and complains of lack of appetite and constipation. Underneath all his words and acts runs the intense egotism and self pity and complaining criticism of those who have tried to help him and self-distaste of his class.

Physical Examination.—He has a markedly neuropathic facies. The eyes are slightly bulging and their expression heavy. The orbicularis oculi muscles are lax and the palpebral fissures sag widely on both sides alike. The pupils are equal and react promptly to light and in accommodation. The visual fields are not cut. The tongue is protruded slightly to the left. The teeth are good and there is no deformity in the mouth. The shape of the head is good and no ill-placement of the ears or any other marked stigma of degeneracy.

The head is held tilted with the chin pointing to the left and the top of the head to the right. While there is present the distinct difference in the size of the two sides of the face there is no loss of function of the facial muscles on either side. The neck: the trapezius on the left is much larger than that on the right (and is hard, giving a suspicion of tumor formation)—a paradoxical hypertrophy. The arm and leg: on the left side they are uniformly smaller than on the right. The bones seem to be of equal size on both sides. The discrepancy is not limited to any muscle or group of muscles. The grip on the left side is good and nearly as strong as on the right. There are no fibrillary twitchings of the muscles on either side of the face, trunk or extremities.

Reflexes.—Knee jerks are prompt and equal; biceps jerk is very prompt on the left and less so on the right; Achilles jerks are equal; triceps jerks are equal; Babinski reflex is absent on both sides; muscle jerks are much more prompt about the shoulder girdle on the left than on the right—this is true of the heavier muscles all over the left side; skin reflexes are more prompt on

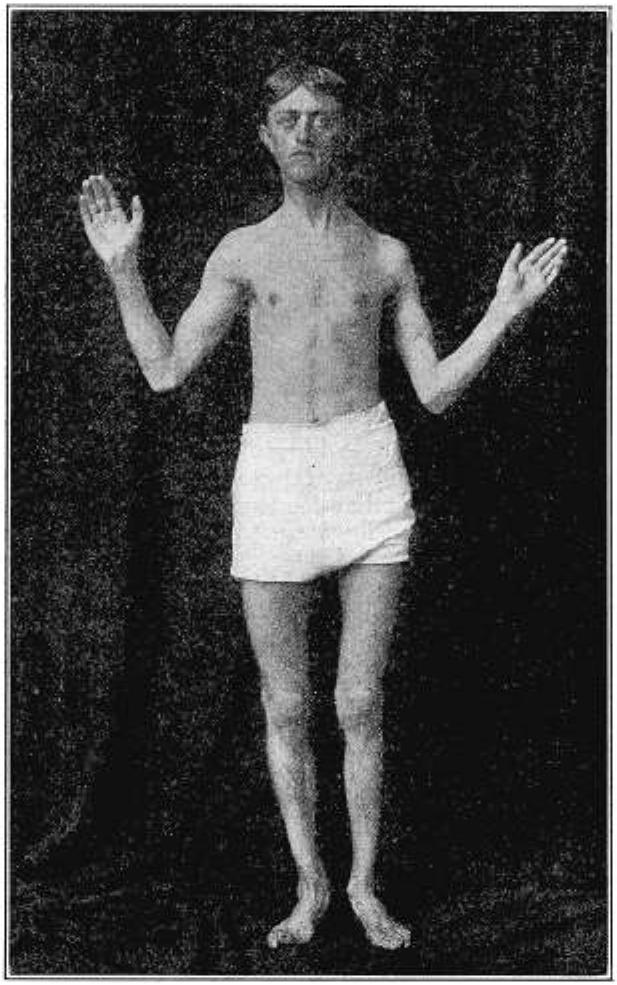

Fig. 1.

the right. There is considerable tilting of the pelvis with consequent spinal deflection that is corrected by raising the right heel five eighths of an inch. Heart and lungs are normal. The abdominal and genital organs are normal.

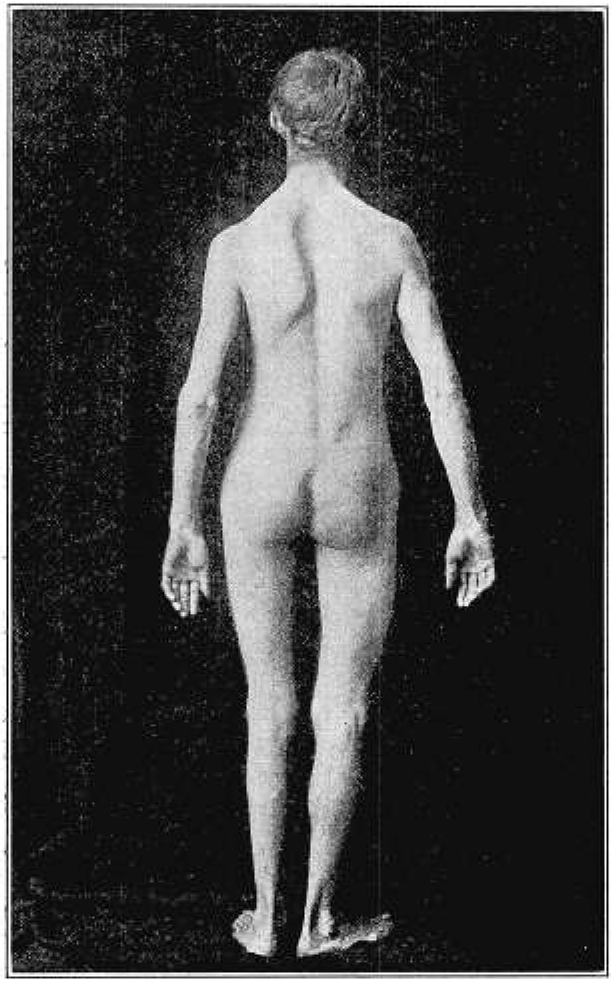

FIG. 2.

Electrical Reactions.—There is no failure to respond to the faradic current on the part of any muscle of the face, arms, legs or thorax. The amplitude is not so great nor the response so prompt on the left side. The reaction time was quicker on the right.

Stigmata of hysteria were not found. The voice is slow, low pitched and listless. There is an evident play for sympathy on the part of the patient that has been taken into account. During the examination he had a "shock": there was no tremor, convulsion or any other outward sign. He was perfectly conscious and talked perfectly connectedly throughout it.

It is noteworthy that he uses the left hand as little as possible: he says he cannot rely on it, and yet his grip is almost as strong on the left as on the right and can be sustained for a long time. The atrophied muscles are not flabby or flaccid.

Sociologically the case is that of a poorly mentally-accoutred, illy-developed, one-sided human inept—to coin a word. Psychologically he would be classed as a malingerer, inasmuch as he consciously and from choice fails to employ what powers he possesses. Neurologically he does not adhere to any one type; there is present the accentuated muscle jerk on the atrophic side that resembles amyotrophic dystrophy, but there is lacking the increased knee jerks, bulbar symptoms and gait. The case is not one of anomaly in which there is present only the failure to develop that happens to be more or less symmetrical: for the differences in the electrical reactions and the skin reflexes together with the marked exaggeration of the muscle jerks on the atrophic side would point to a pathological process, as would also the "shocks" that were felt only on one side. The case is not one merely of asymmetry; the symptom complex would seem to preclude this. The question of asymmetry has been thoroughly worked out. Hunt[12] and Morton,[13] and Roberts,[14] T. Dwight[15] and J. G. Garson[16] have all made reports that correspond closely. They investigated the subject from various points of view. Morton examined over 500 boys and youths and found that about two or three out of every five were asymmetrical, inasmuch as one leg would be shorter than its fellow. They did not report a single case of complete hemiatrophy. In Wight's[17] report of his findings the ratio of asymmetry is placed at one to five.

The case seems to be a trophoneurosis accompanied by a psychosis that may be dependent on it.

As to the probable pathology: "According to de Watteville, trophic changes in muscle may be independent of paralytic phenomena, may affect both muscle and nerve or muscle alone." [18] The trophic, voluntary and reflex influences converge in the multipolar ganglion cell. There may be lesion of one or more of them. If, for example, the trophic and motor influences are

[12] Am. Journ. Med. Sci., January, 1879.

[13] Boston Med. and Surg. Jour., April 15, 1880.

[14] Phila. Med. Times, August 3, 1878.

[15] Mass. Med. Soc. Commu., 1878, p. 125.

[16] Journ. Anat. and Physiol., London, 1878–9, pp. 502–7.

[17] Archi. Clin. Surg., Vol. I, No. 8, February, 1877.

[18] Mills, "The Nervous System and its Diseases," p. 195.

affected there will result a paralysis with atrophy of the muscle. This is the condition in amyotrophic lateral sclerosis. If the lesion is confined to the trophic centers the muscle atrophies but is not paralyzed and presents qualitative alterations in reaction.

It is conceivable that the "unknown agent" of Oppenheim selects the trophic centers for the point of attack. It would seem from the reported cases that the left side, for some reason, is the favorite point.

Mensuration.—The differences in the two sides at various levels are as follows: circumference of head just superior to eyebrows, right half 11.5 in., left half 10.75 in.; of face just inferior to ears, R. 8.5 in., L. 8 in.; of body about the shoulder level: R. 20 in., L. 18.5 in.; about the thorax at nipple level, R. 16 in., L. 14 in.; upper arm, R. 10 in., L. 8.75 in.; thighs, R. 17.5 in., L. 15 in.; calves, R. 12 in., L. 10.5 in.

Conclusions.—The condition is a pathological entity; its beginnings are prenatal in time; the etiological factor is an unknown agent, possibly a toxemia in the fetal circulation that attacks neuropathic individuals and selects the trophic elements, preferably of the left side; the symptoms are atrophy of the muscular structures (or a retardation of their growth); quickening of the muscular reflexes with slowing of the electrical reaction time and without affecting the tendon reflexes materially; asymmetry with accompanying spinal deflection.